# Little Guides Pediatric Melanocytic Lesion Choices

BY BETSY BATES
*Los Angeles Bureau*

MONTEREY, CALIF. — Melanocytic tumors of unknown malignant potential represent some of the most difficult cases in pediatric dermatology, since little agreement exists about their diagnostic criteria, management, or outcome.

"They cause everyone, including pathologists, referring dermatologists, and surgeons, to lose sleep," said Dr. Susan Swetter, director of Stanford (Calif.) University's pigmented lesion and cutaneous melanoma clinic, at the annual meeting of the California Society of Dermatology and Dermatologic Surgery.

Dr. Swetter described the management of an 8-mm raised, blue-black nodule that appeared behind the right ear of an 11-year-old girl. Satellite blue-black macules appeared on the periphery of the lesion.

A partial 5-mm punch biopsy was reviewed by pathologists at Stanford; the University of California, San Francisco; and Massachusetts General Hospital, Boston. The conclusion was that the lesion was a melanocytic tumor of unknown malignant potential (MELTUMP) with decidedly mixed signals: no ulceration but a relatively high mitotic rate ($4/mm^2$) and probable angiolymphatic invasion.

Differential diagnoses included a pigmented epithelioid melanocytoma, an agminated Spitz nevus, or a "low-grade" melanoma (Clark's level III, Breslow thickness 3.7 mm).

The patient underwent a "fairly intuitive" comprehensive work-up, including a thorough personal and family history, a review of the timing and speed of growth of the lesion, and a total body skin examination and physical examination, including palpation of regional lymph node basins to assess for metastasis.

Melanoma experts agree that MELTUMP lesions should be completely excised, but the specifics about recommended margins remain hazy, Dr. Swetter explained. Some experts would decide to perform a wide excision in such a case, perhaps including sentinel lymph node biopsy, as if they were treating a melanoma.

At Stanford, where the patient was seen, the decision was made to take a 1-cm margin, narrower than the 2-cm margin that would be appropriate for a 3.7-mm melanoma, and to await the pathology results before deciding whether to perform a sentinel lymph node biopsy or lymph node dissection.

The histology on the wide excision specimen showed that the lesion was symmetrical and well circumscribed with a polypoid proliferation of darkly pigmented melanocytes and a mitotic rate "well below $1/mm^2$."

Dr. Swetter described the applicable histology images as revealing "deeply pigmented epithelial spindle cells and unmistakable angiolymphatic invasion." "Our pathologists thought this was most consistent with a melanocytoma diagnosis," and noted its rarity as well as its "uncertain biological behavior," said Dr. Swetter.

A comparative genomic hybridization study was ordered from the UCSF laboratory, but results were estimated to take 6-8 weeks, a period of time that could compromise afferent lymphatic drainage from a scalp lesion and reduce the accuracy of the sentinel node biopsy.

After extensive discussions with the child's parents, the Stanford team elected to perform a sentinel lymph node biopsy but to await the outcome of the comparative genomic hybridization studies prior to performing complete lymph node dissection in the event that the sentinel node specimen was positive. A metastatic work-up with PET/CT scanning was performed "in part … to allay some of the parental concern about metastatic disease." Parenthetically, Dr. Swetter noted that such a scan would not generally be indicated in an asymptomatic patient with no signs of metastatic disease and would not preclude the possibility of a positive sentinel node biopsy. The scans were negative.

Two sentinel lymph nodes were identified and removed in the right cervical neck. One was positive for subcapsular and parenchymal metastatic foci of pigmented epithelioid melanocytoma and stained strongly positive for S100, MelanA, and HMB45. MELTUMP lesions have been associated with a very high rate of sentinel lymph node positivity in the two largest retrospective studies to date (44%-50%, compared with about 20% for typical melanomas with Breslow thickness greater than 1 mm.)

However, the picture is confusing, be-

cause studies also associate atypical Spitz tumors with a very high survival rate despite apparent micrometastases.

"The question is, What is the clinical significance [of the high rate of positive sentinel lymph nodes in these tumors]? Does it imply a fatal outcome?" she asked.

In the case of Dr. Swetter's patient, a comparative genomic hybridization offered what appeared to be optimistic information, since the lesion contained aberrations on chromosome 11, a finding that has been exclusively associated with Spitz nevi in comparative studies with other benign nevi and melanomas. The patient has been followed clinically for more than a year without evidence of recurrent disease.

Much uncertainty exists around the management of MELTUMPs; which are characterized by a lack of agreement among pathologists, even those specializing in melanoma pathology.

The lesions should be completely excised, and treated similarly to melanomas when they are characterized by frank atypia or uncertain biologic behavior.

Until more data can be gathered from the national pediatric melanoma and melanocytic neoplasms, Dr. Swetter urged frank discussions among medical professionals and families about the diagnostic uncertainty regarding these lesions. ■

STANFORD UNIVERSITY DEPARTMENTS OF DERMATOLOGY AND PATHOLOGY AND THE MELANOMA CARE COALITION, PHARMADURA, LLC

**The melanocytic tumor of unknown malignant potential on the 11-year-old girl is shown here.**